General

Guideline Title

Os odontoideum. In: Guidelines for the management of acute cervical spine and spinal cord injuries.

Bibliographic Source(s)

Rozzelle CJ, Aarabi B, Dhall SS, Gelb DE, Hurlbert RJ, Ryken TC, Theodore N, Walters BC, Hadley MN. Os odontoideum. In: Guidelines for the management of acute cervical spine and spinal cord injuries. Neurosurgery. 2013 Mar;72(Suppl 2):159-69. [41 references] PubMed

Guideline Status

This is the current release of the guideline.

Recommendations

Major Recommendations

The rating schemes used for the strength of the evidence (Class I-III) and the levels of recommendations (Level I-III) are defined at the end of the "Major Recommendations" field.

Recommendations

Diagnosis

Level III

Plain radiographs of the cervical spine (anterior-posterior, open mouth-odontoid, and lateral) and plain dynamic lateral radiographs performed in flexion and extension are recommended to diagnose and evaluate os odontoideum, with or without tomography (computerized [CT] or plain) and/or magnetic resonance imaging (MRI) of the craniocervical junction.

Management

Level III

- Clinical and radiographic surveillance or posterior C1-C2 internal fixation and fusion are recommended for patients with os odontoideum without symptoms or neurological signs.
- Posterior C1-C2 internal fixation and fusion are recommended for patients with os odontoideum with neurological symptoms, signs, or C1-C2 instability.
- Postoperative halo immobilization is recommended as an adjunct to posterior internal fixation and fusion unless rigid C1-C2 internal fixation is accomplished.
- Occipital-cervical internal fixation and fusion with or without C1 laminectomy is recommended in patients with os odontoideum who have

irreducible dorsal cervicomedullary compression and/or evidence of associated occipital-atlantal instability.

- Ventral decompression should be considered in patients with os odontoideum who have irreducible ventral cervicomedullary compression.

Summary

Plain cervical spine radiographs appear adequate to make a diagnosis of os odontoideum in the vast majority of patients with this disorder. Lateral flexion and extension radiographs can provide useful information regarding C1-C2 instability. Tomography (CT or plain) may be helpful to define the osseous relationships at the skull base, C1, and C2 in patients in whom the craniovertebral junction is not well visualized on plain radiographs. The degree of C1-C2 instability identified on cervical x-rays does not correlate with the presence of myelopathy. A sagittal diameter of the spinal canal at the C1-C2 level of <13 mm does correlate with myelopathy detected on clinical examination. MRI can depict spinal cord compression and signal changes within the cord that correlate with the presence of myelopathy.

Surgical treatment is not required for every patient in whom os odontoideum is identified. Patients who have no neurological deficit and no instability at C1-C2 on flexion and extension studies can be managed without operative intervention. Even patients with documented C1-C2 instability and neurological deficits have been managed nonoperatively without clinical consequence during finite follow-up periods. Most investigators who study and treat this disorder favor operative stabilization and fusion of C1-C2 instability associated with os odontoideum. The concern exists that patients with os odontoideum with C1-C2 instability have an increased likelihood of future spinal cord injury. Although not supported by Class I or Class II medical evidence from the literature, multiple case series (Class III medical evidence) suggest that stabilization and fusion of C1-C2 is meritorious in this circumstance. Because a patient with an initially stable os odontoideum has been reported to develop delayed C1-C2 instability and because there are examples of patients with untreated stable os odontoideum who have developed neurological deficits following minor trauma, surgical consideration and longitudinal clinical and radiographic surveillance of patients with os odontoideum without instability are recommended.

Posterior C1-C2 internal fixation with arthrodesis in the treatment of os odontoideum provides effective stabilization of the atlantoaxial joint in the majority of patients. Posterior wiring and fusion techniques supplemented with postoperative halo immobilization provided successful fusion in 40% to 100% of cases reported. Rigid internal screw fixation and fusion appear to have merit in the treatment of C1-C2 instability in association with os odontoideum and appear to obviate the need for postoperative halo immobilization. Neural compression in association with os odontoideum has been treated with a reduction of deformity, dorsal decompression of irreducible deformity, and ventral decompression of irreducible deformity, each in conjunction with C1-C2 or occipital cervical fusion with internal fixation. Each of these combined approaches has provided satisfactory results. Odontoid screw fixation has no role in the treatment of this disorder.

Definitions:

Rating Scheme for the Strength of the Evidence: Modified North American Spine Society Schema to Conform to Neurosurgical Criteria as Previously Published and for Ease of Understanding and Implementation: Levels of Evidence for Primary Research Question[a]

Class	Therapeutic Studies: Investigating the Results of Treatment	Diagnostic Studies: Investigating a Diagnostic Test	Clinical Assessment: Studies of Reliability and Validity of Observations, Including Clinical Examination, Imaging Results, and Classifications
I	High-quality randomized controlled trial with statistically significant difference or no statistically significant difference but narrow confidence intervals	Testing of previously developed diagnostic criteria on consecutive patients (with universally applied reference "gold" standard)	Evidence provided by 1 or more well-designed clinical studies in which interobserver and intraobserver reliability is represented by a Ä, statistic ≥0.60 or an intraclass correlation coefficient of ≥0.70
	Systematic review[b] of Class I randomized controlled trials (and study results were homogeneous[c])	Systematic review[b] of Class I studies	
II	Lesser-quality randomized controlled trial (e.g., <80% follow-up, no blinding, or improper randomization)	Development of diagnostic criteria on consecutive patients (with universally applied reference "gold" standard)	Evidence provided by 1 or more well-designed clinical studies in which interobserver and intraobserver reliability is represented by a Ä, statistic of 0.40–0.60 or an intraclass correlation coefficient of 0.50–0.70
	Prospective[d] comparative study[e]	Systematic review[b] of Class II studies	
	Systematic review[b] of Class II studies or Class I studies with inconsistent results	Study of nonconsecutive patients; without consistently applied reference "gold"	

Class	Therapeutic Studies: Investigating the Results of Treatment	Diagnostic Studies: Investigating a Diagnostic Test	Clinical Assessment: Studies of Reliability and Validity of Observations, Including Clinical Examination, Imaging Results, and Classifications
	Case-control study[g]	standard Systematic review[b] of Class III studies	
	Retrospective[f] comparative study[e]	Case-control study	
	Systematic review[b] of Class II studies		
III	Case series[h]	Poor reference standard	Evidence provided by 1 or more well-designed clinical studies in which interobserver and intraobserver reliability is represented by a Ä, statistic of <0.40 or an intraclass correlation coefficient of <0.50
	Expert opinion	Expert opinion	

[a]A complete assessment of quality of individual studies requires critical appraisal of all aspects of the study design.

[b]A combination of results from 2 or more prior studies.

[c]Studies provided consistent results.

[d]Study was started before the first patient enrolled.

[e]Patients treated 1 way (e.g., halo vest orthosis) compared with a group of patients treated in another way (e.g., internal fixation) at the same institution.

[f]The study was started after the first patient was enrolled.

[g]Patients identified for the study on the basis of their outcome, called "cases" (e.g., failed fusion), are compared with those who did not have outcome, called "controls" (e.g., successful fusion).

[h]Patients treated 1 way with no comparison group of patients treated in another way.

Levels of Recommendation

Level I	Generally accepted principles for patient management, which reflect a high degree of clinical certainty (usually this requires Class I evidence which directly addresses the clinical questions or overwhelming Class II evidence when circumstances preclude randomized clinical trials)
Level II	Recommendations for patient management which reflect moderate clinical certainty (usually this requires Class II evidence or a strong consensus of Class III evidence)
Level III	Other strategies for patient management for which the clinical utility is uncertain (inconclusive or conflicting evidence or opinion)

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

Os odontoideum

Guideline Category

Diagnosis

Evaluation

Management

Treatment

Clinical Specialty

Neurological Surgery

Orthopedic Surgery

Pediatrics

Radiology

Intended Users

Advanced Practice Nurses

Hospitals

Nurses

Physician Assistants

Physicians

Guideline Objective(s)

To update the medical evidence on the diagnosis and management of patients with os odontoideum since the 2002 publication

Target Population

All patients with os odontoideum, including:

- Patients with occipital-cervical pain alone
- Patients with myelopathy
- Patients with intracranial symptoms or signs from vertebrobasilar ischemia

Interventions and Practices Considered

Diagnosis/Evaluation

1. Plain radiographs of the cervical spine (anterior-posterior, open mouth-odontoid, and lateral)
2. Plain dynamic lateral radiographs performed in flexion and extension
3. Tomography (computerized [CT] or plain) of the craniocervical junction
4. Magnetic resonance imaging (MRI) of the craniocervical junction

Treatment/Management

1. Clinical and radiographic surveillance
2. Posterior C1-C2 internal fixation and fusion
3. Postoperative halo immobilization
4. Occipital-cervical internal fixation and fusion with or without C1 laminectomy
5. Ventral decompression

Major Outcomes Considered

- Utility of imaging studies in establishing diagnosis
- Radiographic stability (fusion rate)
- Complications of surgery

Methodology

Methods Used to Collect/Select the Evidence

Hand-searches of Published Literature (Primary Sources)

Hand-searches of Published Literature (Secondary Sources)

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

Search Criteria

A National Library of Medicine (PubMed) computerized literature search from 1966 to 2011 was performed through MEDLINE using the key phrase "os odontoideum." The search identified 224 articles. Articles written in English were reviewed. Thirty-eight articles that described the clinical aspects and management of patients with os odontoideum were identified and used to generate these guidelines. None of the articles meeting selection criteria provided Class I or Class II medical evidence. All 38 citations offered Class III medical evidence on the diagnosis and/or management of os odontoideum.

Number of Source Documents

Thirty-eight articles represent the basis for this review and are summarized in Evidentiary Table format (see the table in the original guideline document).

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Rating Scheme for the Strength of the Evidence: Modified North American Spine Society Schema to Conform to Neurosurgical Criteria as Previously Published and for Ease of Understanding and Implementation: Levels of Evidence for Primary Research Question[a]

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Class	Therapeutic Studies: Investigating the Results of Treatment	Diagnostic Studies: Investigating a Diagnostic Test	Clinical Assessment: Studies of Reliability and Validity of Observations, Including Clinical Examination, Imaging Results, and Classifications
	blinding, or improper randomization)	(with universally applied reference "gold" standard)	reliability is represented by a Ä statistic of 0.40–0.60 or an intraclass correlation coefficient of 0.50–0.70
	Prospective[d] comparative study[e]	Systematic review[b] of Class II studies	
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Methods Used to Analyze the Evidence

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

Selected articles were carefully reviewed by the authors. An evidentiary table was created (refer to the table in the original guideline document) that reflected the strengths and weaknesses of each article.

On occasion, the assessed quality of the study design was so contentious and the conclusions so uncertain that the guideline authors assigned a lower medical evidence classification than might have been expected without such a detailed review. In every way, adherence to the Institute of Medicine's criteria for searching, assembling, evaluating, and weighing the available medical evidence and linking it to the strength of the recommendations presented in this document was carried out.

Articles that did not achieve immediate consensus among the author group were discussed extensively until a consensus was reached. Very few contributions required extensive discussion. Most articles were easily designated as containing Class I, II, or III medical evidence using the criteria set forth by the author group at the initiation of the literature evaluation process (see the "Rating Scheme for the Strength of the Evidence" field).

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

The current author group was selected for its expertise in spinal surgery (both neurosurgical and orthopedic), neurotrauma, clinical epidemiology, and, in several cases, prior experience with guideline development. The topics chosen for inclusion in this iteration of these guidelines are contemporary and pertinent to the assessment, evaluation, care, and treatment of patients with acute cervical spine and/or spinal cord injuries.

Rating Scheme for the Strength of the Recommendations

Levels of Recommendation

Level I	Generally accepted principles for patient management, which reflect a high degree of clinical certainty (usually this requires Class I evidence which directly addresses the clinical questions or overwhelming Class II evidence when circumstances preclude randomized clinical trials)
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Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

Not stated

Description of Method of Guideline Validation

Not applicable

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see the "Major Recommendations" field). All articles provided Class III medical evidence.

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Accurate diagnosis and appropriate management of os odontoideum

Potential Harms

Reported complications of fixation and fusion techniques include:

- Wound infection

- Prolonged dysphagia
- New neurological deficits
- Vertebral artery injuries
- Cerebrospinal fluid leak

Qualifying Statements

Qualifying Statements

- Medical evidence-based guidelines are not meant to be restrictive or to limit a clinician's practice. They chronicle multiple successful treatment options (for example) and stratify the more successful and the less successful strategies based on scientific merit. They are not absolute, "must be followed" rules. This process may identify the most valid and reliable imaging strategy for a given injury, for example, but because of regional or institutional resources, or patient co-morbidity, that particular imaging strategy may not be possible for a patient with that injury. Alternative acceptable imaging options may be more practical or applicable in this hypothetical circumstance.
- Guidelines documents are not tools to be used by external agencies to measure or control the care provided by clinicians. They are not medical-legal instruments or a "set of certainties" that must be followed in the assessment or treatment of the individual pathology in the individual patients we treat. While a powerful and comprehensive resource tool, guidelines and the recommendations contained therein do not necessarily represent "the answer" for the medical and surgical dilemmas faced with many patients.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Mobile Device Resources

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

IOM Domain

Effectiveness

Identifying Information and Availability

Bibliographic Source(s)

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Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2013 Mar

Guideline Developer(s)

American Association of Neurological Surgeons - Medical Specialty Society

Congress of Neurological Surgeons - Professional Association

Source(s) of Funding

Congress of Neurological Surgeons

Guideline Committee

Guidelines Author Group of the Joint Section of Disorders of the Spine and Peripheral Nerves of the American Association of Neurological Surgeons and the Congress of Neurological Surgeons

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Financial Disclosures/Conflicts of Interest

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this guideline.

Guideline Status

This is the current release of the guideline.

Guideline Availability

Electronic copies: Available in Portable Document Format (PDF) and EPUB for eBook devices from the Neurosurgery Web site .

Availability of Companion Documents

The following are available:

- Foreword. Guidelines for the management of acute cervical spine and spinal cord injuries. Neurosurgery 2013;72(3):1. Electronic copies: Available in Portable Document Format (PDF) from the Neurosurgery Web site .
- Commentary. Guidelines for the management of acute cervical spine and spinal cord injuries. Neurosurgery 2013;72(3):2-3. Electronic copies: Available in PDF from the Neurosurgery Web site .
- Introduction to the guidelines for the management of acute cervical spine and spinal cord injuries. Neurosurgery 2013;72(3):5-16. Electronic copies: Available in PDF from the Neurosurgery Web site .
- Methodology of the guidelines for management of acute cervical spine and spinal cord injuries. Neurosurgery 2013;72(3):17-21. Electronic copies: Available in PDF from the Neurosurgery Web site .

Patient Resources

None available

NGC Status

This NGC summary was completed by ECRI Institute on July 9, 2013. The information was verified by the guideline developer on October 3, 2013.

Copyright Statement

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